Renal surgery in patients with a duplicated inferior vena cava: a case series and review of the literature

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Abnormal inferior vena cava (IVC) anatomy may present unique challenges for urologists when performing retroperitoneal surgery. Duplication of the IVC is one such anomalous variation and can be found in up to 3% of the population. Misunderstanding of the implications of this aberrant anatomy may lead to intraoperative or postoperative complications. Here, we present two cases of patients undergoing renal surgeries with duplicate IVC. We then review the embryologic origin and anatomic findings in those with abnormal IVC anatomy as well as discuss the surgical implications and considerations for urologists.

Key Words: inferior vena cava, anatomical variation, urologic surgery, renal

Introduction

Anatomical variations present unique challenges for surgeons and are often cited as reasons for intraoperative errors.[1] Anomalies of the inferior vena cava (IVC) and renal vasculature are common and have a prevalence of over five percent.[2] Duplication of the inferior vena cava (DIVC) may occur in between 0.2% and 3% of the population.[3] These vasculature aberrations can create unique challenges for urologists when performing renal or retroperitoneal surgery. Here, we present a series of two patients with DIVC who underwent surgery at our institution. This is followed by a discussion of surgical implications to consider when operating on this unique patient population.

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Case 1

A 57-year-old male with a family history of bilateral multifocal tumors and a negative genetic work up presented after a diagnosis of a 4.5 cm left lower pole mass on screening renal ultrasound. He had no past surgical history. Preoperative creatine clearance was 92 milliliter (mL)/minute (min). He underwent dedicated cross sectional imaging studies which noted a DIVC and confirmed the diagnosis of a renal mass, Figure 1. He was subsequently taken to the OR for a robot-assisted laparoscopic left partial nephrectomy. Perioperative heparin was given subcutaneously. Intraoperatively, a DIVC with associated duplicated renal vein was visualized, Figure 2. Only one gonadal vein was identified, which drained into the larger of the two renal veins (patient also had two renal arteries). The larger renal vein drained into the right sided IVC, and the diminutive duplicate renal vein did not provide significant venous drainage of the kidney but drained into the left sided IVC. This smaller renal vein was ligated intraoperatively so that a window superior to the larger renal vein could be exposed for potential hilar clamping. During the case the partial nephrectomy was performed while clamping both renal arteries for a clamp time of 23 minutes. There were no intraoperative or postoperative complications. Pathology was consistent with smooth muscle rich clear cell renal cell carcinoma. Postoperative creatinine clearance two months later was 85 mL/min.

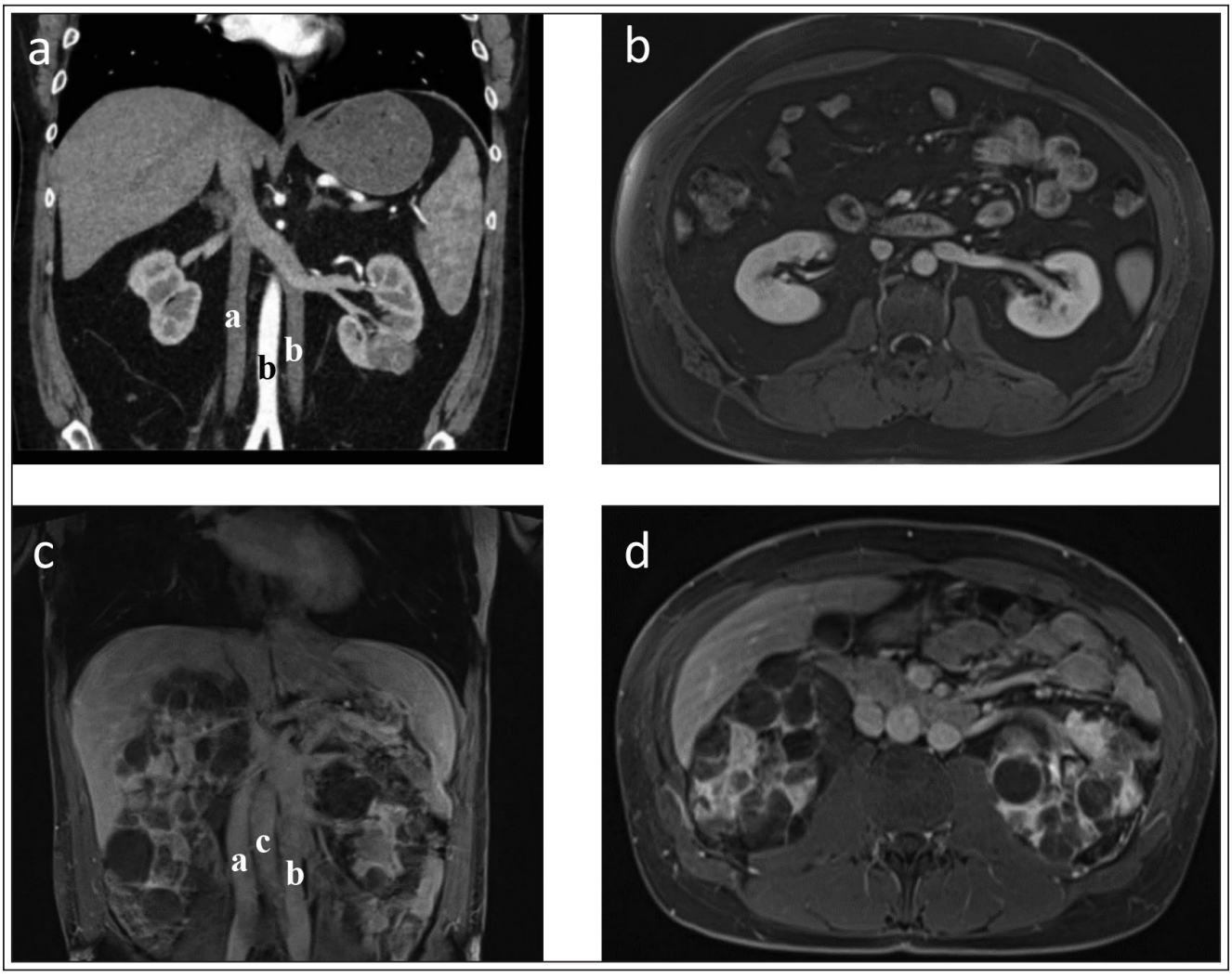

Figure 1. (a) case one coronal view; **(b)** axial view with left renal vein extending from duplicated IVC; **(c)** case two coronal view; **(d)** case two axial view showing DIVC. Index: a = right-sided IVC; b = left-sided IVC; c = aorta

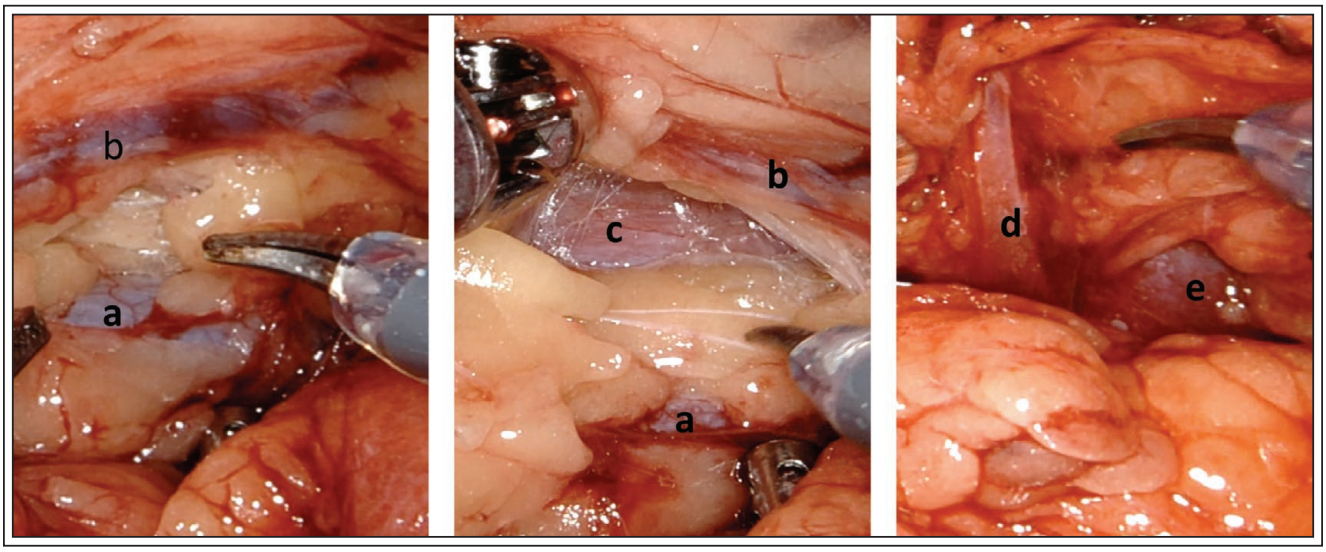

Figure 2. Intraoperative images from Case 1. Index: a = left IVC; b = left gonadal vein; c = psoas muscle; d = left duplicate renal vein (draining into left IVC); e = left main renal vein draining into right IVC.

Case 2

A 39-year-old male with a diagnosis of anemia, chronic kidney disease and von Hippel-Lindau (VHL) syndrome with several prior renal masses presented with multiple, large bilateral renal masses. His prior renal procedures had included a left robotic partial nephrectomy with removal of 73 lesions (11 consistent with renal cell carcinoma), prior right radiofrequency ablation and partial nephrectomy, and bilateral renal cyberknife therapy. Preoperative creatine clearance was 31 mL/min. Cross sectional imaging was similarly notable for a DIVC with three renal arteries on each side, Figure 1. Given his extensive prior surgical history, poor renal function, and bilateral renal masses measuring over 3.5 cm on the right and 3.8 cm on the left with extensive mixed cystic/solid lesions encompassing the majority of both kidneys, the decision was made to undergo bilateral open nephrectomy. A bilateral subcostal (Chevron) incision was made. The retroperitoneum was exposed by completely mobilizing the right colon and small bowels to the Ligament of Treitz and to best visualize the DIVC and bilateral renal veins, which were single and draining into their respective IVCs, Figure 3. The right gonadal vein was seen coursing into the right IVC, whereas the left gonadal vein drained into the left renal vein, which also emptied the left sided IVC. The renal vein was ligated laterally to the insertion of the left IVC, sparing this structure. There were no intraoperative complications. Postoperatively, the patient received two units packed red blood cells but otherwise started hemodialysis without issue. Pathology was consistent with bilateral clear cell renal cell carcinoma grade 2 throughout the entirety of both kidneys.

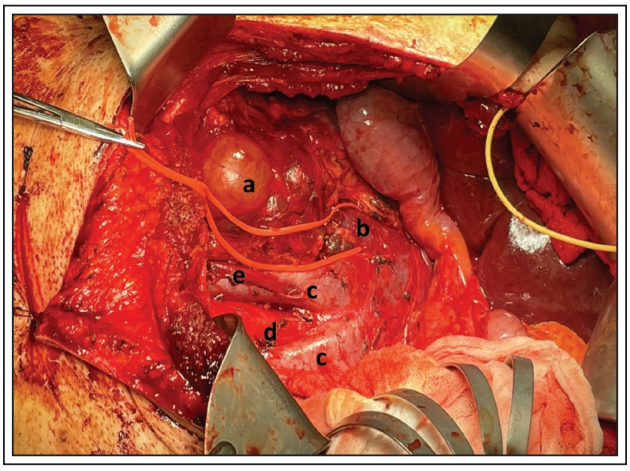

Figure 3. Intraoperative images from Case 2. Index: a = renal cyst; b = right renal vein; c = right IVC; d = aorta; e = right gonadal vein.

Discussion

DIVC presents a unique challenge for surgeons, particularly urologists in the retroperitoneum. It may complicate or prolong urologic surgery, particularly

increasing the challenge for left sided cases, where the variant anatomy may confuse urologists and present an increased opportunity for intraoperative complications including mistaking the duplicated IVC as a gonadal vein. An understanding of the anatomic variances associated with DIVC is crucial to avoid vascular injury or postoperative complications.

The IVC is thought to form after the sixth to seventh week of gestation. Initially, three paired venous drainage structures exist, including the posterior cardinal veins, the subcardinal veins, and the supracardinal veins. It is the unilateral regression of these paired structures that result in the formation of the IVC. The posterior cardinal veins account for the distal IVC, whereas the supracardinal and subcardinal veins join to become the infrarenal and supraprenal IVC, respectively. The anastomosis of these right sided veins with the concurrent regression of their left sided counterparts results in a right sided IVC. Abnormalities with this embryologic process may be associated with horseshoe kidney and several IVC aberrant anatomies.[4]

DIVC results from the persistence of the infrarenal segment of the left sided embryologic supracardinal vein. In most instances this results in a left sided IVC draining into the left renal vein, which normally drains into an orthotopic right IVC. Similarly, the iliac drainage is typically into the respective IVCs. Some cases may have an interiliac connection, providing an important source of venous collateral flow in those with DIVC. Prior series have noted that while the ipsilateral gonadal and lumbar veins typically drain into the ipsilateral IVC, they may also drain into the renal vein on the left side.[5] While less commonly reported in the literature with cases of DIVC, we suspect that renal vasculature anomalies may be present at similar or higher rates as those without IVC anomalies.

While a DIVC may result from failure of regression of the left sided venous structures, several other morphologic variants may also form from aberrant embryogenesis. Most commonly, DIVC or a solitary left IVC, known as transposition of the IVC, are seen, however; circumcaval ureters and azygous continuation of the IVC may be observed.[6] Importantly, a circumcaval ureter, resulting from persistence of the right posterior cardinal vein and regression of the right supracardinal vein, may cause impingement of the ureter, which passes posteriorly to the right IVC in these cases resulting in severe hydronephrosis. We could find no significant association between abnormalities in IVC development and renal artery or ureteral anatomy anomalies, as expected given the separate embryologic origin of these structures.[6]

The diagnosis of DIVC is made radiographically, and most are found incidentally on cross sectional computed tomography (CT) images. While usually evident on CT imaging, contrast enhancement, with a slight delay after injection, may allow IVC enhancement for better diagnosis and to evaluate for occlusion or intraluminal masses. In cases where the diagnosis is unclear or CT is contraindicated, magnetic resonance (MR) imaging or doppler ultrasound may be used, though the latter may be less specific and sensitive in many patient populations.[7] MR or CT venography may allow clinicians to better assess for venous collateralization in the cases of extirpative surgeries.

DIVC requires significant preoperative planning and evaluation on the part of the urologist in order to avoid renal injury or devascularization of renal segments. In patients with DIVC, the left renal vein typically inserts into the left IVC. However, duplicate renal veins may be present with insertions on both the left IVC and orthotopic right IVC. Similarly, the gonadal vasculature may be aberrant, with insertion into the left renal vein or left IVC. While the left gonadal vein hemodynamics portend a higher risk of varicocele on the left side already, surgical ligation of the gonadal vein may predispose patients to symptomatic scrotal edema or hydroceles.[8] Similarly, there have been cases of thigh and pelvic girdle edema following ligation of the left infrarenal vena cava, thought to be due to impaired venous return and insufficient venous collateralization draining the ipsilateral lower extremity.[9]

Little is known about any associations with IVC congenital malformations and the risk of venous thromboembolism (VTE), with most of the data on DIVC and IVC malformations presented in the form of small case reports or case series. However, some studies have demonstrated a higher rate of venous thromboembolism in patients with IVC malformations.[10,11] Some authors have argued that DIVC may be associated with an increased risk of venous stasis, thereby predisposing patients to VTE.[11] Interestingly, in patients with VTE events and IVC malformations, bilateral deep venous thrombosis is thought to occur at a higher rate, supporting the theory that these patients may be at higher risk of venous stasis. While we could find no data to support the routine use of VTE chemoprophylaxis in patients with DIVC undergoing urologic surgery, our review of the literature suggests that VTE prophylaxis should strongly be considered. When VTE events occur and therapeutic anticoagulation is contraindicated, IVC filters may need to be placed in both the left and right sided IVC as clot may embolize via either vessel.

Renal cell carcinoma with extension of tumor thrombus into the IVC represents a significant challenge for many urologists, and surgery may be associated with significant vascular morbidity including hemorrhage or VTE. In some instances, tumor thrombus may be invading the IVC itself, requiring removal of a portion of the cava. While there is little data in those with left sided tumors and DIVC, removal of the left IVC may be feasible in certain scenarios. To safely perform this procedure, venous collateralization may be determined preoperatively with CT or MR venogram to prevent postoperative lower extremity edema or pain.[12]

Surgically, broad dissection and visualization of the retroperitoneal vasculature is essential to avoiding vascular injury. Typically, a midline incision or Chevron incision, as opposed to a flank incision, would be preferable for best exposure of the aberrant great vessels. In our open bilateral nephrectomy, we obtained extensive visualization of both the right and left IVC as well as the aorta. The renal arteries and veins were identified at their takeoffs from the aorta and DIVC, respectively. This allowed us to assess for any vascular aberrancies not seen on preoperative cross-sectional imaging. Care was taken to identify the gonadal veins bilaterally as well, so that they could be spared. The left renal vein should be ligated lateral to the insertion of the gonadal vein and IVC, when possible, to prevent postoperative complications. Similarly, in cases such as retroperitoneal lymph node dissections or left sided adrenal surgeries, adequate visualization of the great vessels and aberrant IVC anatomy will allow the surgeon to prevent vascular injury, which may be associated with hemorrhage or lower extremity thrombosis or edema.

In the robotic approach, care must be taken to not confuse the left sided IVC with the gonadal vein, which runs along the same course but is often lateral and of a smaller caliber than the aberrant IVC. When necessary, care must be taken to avoid ligation of any duplicate renal vein in nephron-sparing approaches to prevent subsequent venous congestions or hyperperfusion injury. However, as in the case with our patient, ligation may be necessary in some cases in order to adequately visualize and control the renal hilum.

Conclusions

We present two patients who underwent urologic surgery who were found to have duplication of the inferior vena cava. In cases where aberrant IVC anatomy is suspected, preoperative imaging and diagnosis is crucial to avoid intraoperative surgical complications. There are several aberrant vascular malformations that may occur with DIVC, and care must be taken intraoperatively to avoid venous injury. Patients with DIVC may be more prone to renal abnormalities or VTE events, and strong consideration should be given to perioperative DVT chemoprophylaxis. Despite the aberrant anatomy, patients with DIVC may successfully undergo urologic operations with successful outcomes. □

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